CASE REPORT

Kienbock's disease and juvenile idiopathic arthritis

Nicholas M. Desy*, Mitchell Bernstein, Edward J. Harvey, Elizabeth Hazel

ABSTRACT: Kienbock's disease or osteonecrosis of the lunate is an uncommon cause of wrist pain. . Though there have been several reports of cases in patients with various rheumatologic diseases, the precise etiology has currently not been established. We report a case of Kienbock's disease that occurred in a patient with juvenile idiopathic arthritis. To our knowledge, this is the first case report with an association between these two conditions.

Keywords: Kienbock's disease, osteonecrosis, juvenile idiopathic arthritis, lunatomalacia, avascular necrosis

INTRODUCTION

The etiology of Kienbock's disease, also known as (osteonecrosis of the lunate, remains controversial. It commonly occurs in patients twenty to fourty years old and presents with pain and stiffness in the dorsomedial aspect of the wrist. Several risk factors have been established to help explain its etiology: acute or repetitive trauma, variation in blood supply to the lunate, differences in the anatomy and shape of the lunate bone, and venous congestion (1, 2). Abnormal biomechanics at the radiocarpal joint between the distal radius and ulna has also been implicated in Kienbock's disease (3, 4). Ulnar variance describes the length relationship between the articular surfaces of the radius and ulna at the radiocarpal joint. Positive ulnar variance indicates that the ulna is longer than the radius, while negative ulnar variance indicates that the ulna is shorter at the wrist joint. In neutral ulnar variance 80% of the axial load at the wrist is transmitted through the distal radius. As ulnar variance decreases to more negative values, the load transmission across the distal radiocarpal joint increases, subsequently exposing the lunate to abnormally higher pressures and potentially increasing the risk of Kienbock's disease (3, 4).

Kienbock's disease is also associated with systemic lupus erythematosus (SLE) (5-8), antiphospholipid antibody syndrome (9), sickle cell anemia (10), and Crohn's enteritis (11). Multiple hereditary osteochondromata (12), carpal coalition (13, 14) and congenital shortening of the ulna in Langer-Giedion syndrome (15), are other anatomic abnormalities that have been reported with Kienbock's disease. Rheumatic diseases, including scleroderma (16-18), rheumatoid arthritis (19), gout (20, 21) and dermatomyositis (22) have been published in association with Kienbock's, but there have been no identifiable cases in patients with juvenile idiopathic arthritis (JIA).

This report presents a case of osteonecrosis of the lunate in a patient with JIA and no prior history of trauma. Furthermore, a literature review is done to illustrate the proposed etiologies of Kienbock's disease and its association with other rheumatologic conditions.

*To whom correspondence should be addressed:
Dr. Nicholas M. Desy
McGill University Health Centre Montreal General Hospital
1650 Cedar Avenue Room Montreal, Quebec, Canada, H3G 1A4
Tel: (514) 934-1934 ext. 42734; Fax: (514) 934-8453
Email: nicholas.desy@mail.mcgill.ca

CASE REPORT

A 20-year-old right-handed female with known rheumatoid factor negative polyarticular JIA presented to the clinic because of pain and limited range of motion in the left wrist.

She was diagnosed with JIA at the age of nine after a two-month history of pain and swelling in both hands and knees. She reported difficulty with recreational activities. During the course of her illness several other joints progressively became involved. During the first year of treatment, she was prescribed nonsteroidal anti-inflammatory medication and low-dose prednisolone. To help control her symptoms she required disease-modifying antirheumatic drugs. Her medications included methotrexate 20 mg weekly, folinic acid 2.5 mg weekly, and etanercept 25 mg twice a week.

Three weeks prior to presentation, the patient experienced a severe flare of her arthritis due to non-compliance with her medication. This lead to persistent left wrist pain and limited range-of-motion. On examination she demonstrated synovial thickening of her left wrist with no palpable effusion.

Magnetic resonance imaging of her left wrist showed mild synovial thickening and erosive arthropathy throughout the carpus, radiocarpal, and carpometacarpal joints. In addition, there is possible sclerosis and edema, since she had collapse of the lunate with mixed signal intensity. Plain radiographs revealed negative ulnar variance, sclerosis, and loss of lunate height (Fig. 1). The imaging was compatible with Stage 4 osteonecrosis of the lunate (23). The patient was managed non-operatively and at two-year follow-up was asymptomatic with no concomitant worsening of lunate osteonecrosis on radiograph.

DISCUSSION

The etiology of Kienbock's disease remains unclear (Table 1). The current literature indicates that most cases of Kienbock's disease develop without a history of trauma and posit that e extraosseous and intraosseous blood supply to the lunate have a role in the disease process. The extraosseous blood supply is formed by a series of dorsal and volar vascular arches (1, 24). The intraosseous blood supply is made up of braches entering the volar and dorsal poles however the composite of this vasculature is variable (1, 25-27); branches demonstrate different anastomosis patterns inside the lunate, or the lunate may be supplied by only one dorsal or volar branch or By both a dorsal and volar arterial supply the lunate without any anastomosis (26). When an anastomosis does exist, it can be characterized as a Y, X, or I pattern, depending on the amount of vessels supplying each pole (24, 27). Depending on the intraosseous vascular anatomy

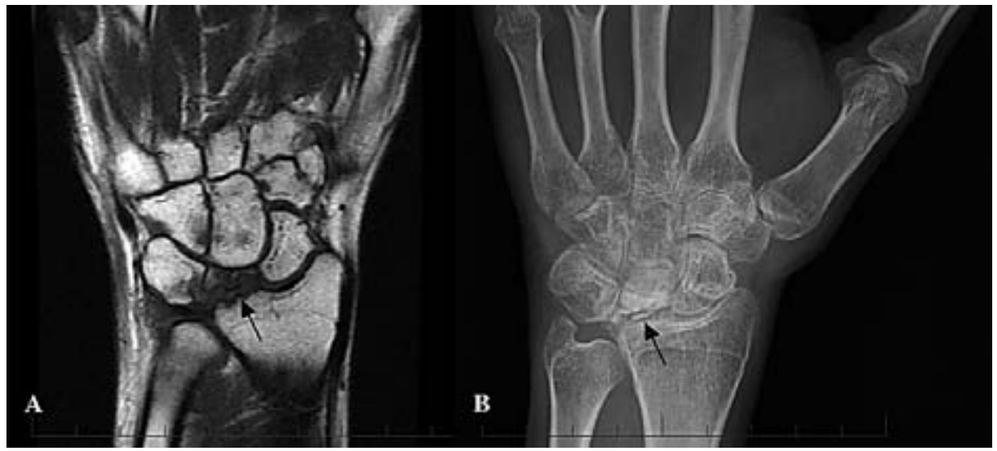

Figure 1: A. Coronal T1-weighted fast-spin-echo magnetic resonance image showing collapse of the lunate with mixed signal intensity suggesting sclerosis and edema consistent with Kienbock's disease (arrow). Erosive arthropathy throughout the carpus and negative ulnar variance are also noted. B. Antero-posterior plain X-ray of the left wrist demonstrates sclerosis and slight loss of height involving the lunate compatible with Kienbock's disease (arrow). Negative ulnar variance and degenerative changes are also seen.

Current proposed mechanisms
Aberrant blood supply to the lunate
Abnormal risk biomechanics
Endothelial cell dysfunction
Increased intraosseous pressure
Microvascular thrombophilia
Trauma-induced with disruption of the blood supply
Venous congestion

Table 1: Pathogenesis of osteonecrosis of the lunate

of the lunate, certain lunate bones are predisposed to Kienbock's disease. This concept was highlighted in a case report of Kienbock's disease associated with sickle cell anemia (10). The osteonecrosis was thought to have developed from an at-risk lunate - single volar arterial supply - along with significant vascular sickling and stasis.

Venous congestion has also been attributed to the pathogenesis of Kienbock's disease (2). During surgery, Jensen measured increased pressure inside the lunate compared with the radial styloid and capitate. He concluded that the higher pressure was caused by venous congestion leading to osteonecrosis of the lunate.

Negative ulnar variance is also implicated in the pathogenesis of Kienbock's disease (3, 4, 28). The altered relationship between the ulna and radius at the distal radioulnar joint modifies the biomechanics at the radiolunate joint and increases strains on the lunate. This postulation is still controversial because several studies, including a meta-analysis, have shown that negative ulnar variance is not a risk factor for developing Kienbock's disease, (29, 30). On the contrary,Ledoux et al. performed a finite-element analysis on cadaveric lunate bones and found that the progression of a fracture of the lunate was present with negative ulnar variance, a high lunate uncovering index, which is the amount of lunate outside the lunate fossa of the radius compared to the amount of lunate articulating with the lunate fossa, and angulated trabeculae (31). This suggests that given the circumstance, the lunate can be at risk for developing osteonecrosis due to abnormal stresses.

Further cases have also reportedpatients with conditions that may have caused altered stresses on the lunate. In particular, two cases have been reported involving carpal coalition (13, 14). It was postulated in these cases that carpal coalition caused a progressively increasing stress on the lunate, which in turn led to Kienbock's disease. Schuind et al. reported a case of Kienbock's disease associated with congenital shortening of the ulna as seen in Langer-Giedion syndrome (15). It was suggested that Kienbock's disease developed from microfractures sustained by an abnormal stress distribution (15). Multiple hereditary osteochondromata in the forearm was also found in association with Kienbock's disease and was attributed to an excess load on the lunate by negative ulnar variance, but with no carpal slip (12).

Systemic lupus erythematosus has been associated with avascular necrosis of bone. In 1977, Urman presented several cases of patients with SLE and osteonecrosis of the carpal bones, including a case report of a patient with SLE and Kienbock's disease (8). The patient also had a history of Raynaud's phenomenon and was taking high-dose corticosteroids. In SLE patients treated with corticosteroids and who developed osteonecrosis, there was one patient who developed Kienbock's disease (5). This patient was treated with large doses of corticosteroids compared to those who did not develop Kienbock's disease. Mok et al. reported a case of bilateral Kienbock's disease in a patient with SLE (6). The patient described in this case report was never treated with corticosteroids and the etiology of Kienbock's disease was thought to be due to a vasculopathy caused by either vasculitis or antiphospholipid antibodies, even though this patient tested negative for lupus anticoagulant and anticardiolipin antibodies. More recently, Taniguchi et al. described two cases of Kienbock's disease in SLE after taking high doses of steroids (7). One of the cases was of a patient who developed bilateral osteonecrosis of the lunate. Both cases attributed corticosteroid use with the development of osteonecrosis. It is apparent that the cases of Kienbock's disease in patients with SLE were either attributed to high dose corticosteroid use or to the disease itself. More over, patients with Crohn's disease who use corticosteroids to control disease symptoms are known to develop Kienbock's disease and osteonecrosis of the hip (11). The same scenario also occurred in a patient with dermatomyositis taking high doses of corticosteroids for sixteen months (22).

Scleroderma associated with Kienbock's disease was first reported by Agus et al. in a patient with bilateral osteonecrosis of the lunate (16). This patient had severe Raynaud's phenomenon and was never treated with corticosteroids. The contributing factors were hypothesized to be vasculopathy, Raynaud's phenomenon, and a lunate consisting of a single nutrient vessel. Ribbans also reported a case of Kienbock's disease in a patient with scleroderma and severe Raynaud's phenomenon (18). The vasculopathy, scleroderma, and repeated use of the patient's affected wrist predisposed this patient to Kienbock's disease. Matsumoto et al. reported three cases of Kienbock's disease in three patients with scleroderma, two without a history of steroid use and one who only used low dose steroids prior to the diagnosis of osteonecrosis (17). All three patients also had limited skin involvement, but had severe Raynaud's phenomenon. They reported that scleroderma related vascular disease was likely the cause of the circulatory impairment leading to osteonecrosis.

Mok et al. reported a patient with rheumatoid arthritis who was found to have osteonecrosis of the lunate. They believed that the Kienbock's disease occurred in this patient due to an increase in intra-articular pressure within the wrist compartment, causing impedance of venous return, vascular insufficiency to the lunate, and subsequent osteonecrosis (19).

In addition to the various risk factors mentioned above, Kienbock's disease b is prevalent in specific patient populations. Rooker et al. found an increased prevalence of Kienbock's disease in a group of patients with cerebral palsy (9.4%) (32). This was thought to be related to an abnormally flexed posture of a spastic wrist, which was present in all patients with Kienbock's disease in this study and could impede blood flow. Joji et al. also found an increased prevalence of Kienbock's disease in patients with cerebral palsy (2.7%) (33). They believed that the high muscle tone across the wrist in an ulnar negative wrist caused an increased pressure on the lunate. This cause is consistent with repeated microtrauma leading to vascular compromise and ultimately osteonecrosis.

While the association between Kienbock's disease and several of the reported cases could be coincidental, having a rheumatologic co-morbidity such as JIA could represent a risk factor for Kienbock's disease, as seen in the presented case report. The wrist is the second most common site of growth abnormality in patients with JIA (34). Several changes occur at the wrist, including narrowing of the intercarpal spaces, premature ossification of the carpal bones, and early fusion of the ulnar epiphysis leading to a shorter ulna (negative ulnar variance) (35). The wrist ultimately becomes displaced ulnarly and volarly leading to a dislocation of the wrist and bayonet deformity. Therefore, it is possible that the abnormalities in the wrist associated with JIA could lead to abnormal stresses and or pressures in the wrist that led to Kienbock's disease. Furthermore, the erosive changes in other carpal bones may lead to a change on the normal force patterns in the patient's carpus. Seven years prior to the onset of Kienbock's disease, our patient was also treated with low-dose corticosteroids which could have also disrupted circulation and led to the development of lunate osteonecrosis.

The precise etiology of Kienbock's disease remains elusive. Several theories attempt to explain its pathogenesis, which suggests that it may be multifactorial. Many risk factors have also been identified; steroid treatment, a predisposing rheumatologic disease, a variation in lunate blood supply, and possibly negative ulnar variance. Our case demonstrates the possible relation between Kienbock's disease and JIA. It also suggests that Kienbock's disease could be a possible cause of wrist pain and stiffness in patients with JIA.

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Nicholas M. Desy, M.D.C.M. is a third year resident in Orthopaedic Surgery at McGill University. He obtained a B.Sc. in Microbiology and Immunology from McGill University. He then completed his medical degree in 2008 from McGill University.

Mitchell Bernstein, M.D. is a fourth year resident in Orthopaedic Surgery at McGill University. He received a B.Sc. in Microbiology and Immunology from McGill University followed by an M.D. in Chicago.

Edward J. Harvey M.D., M.Sc. is an Associate Professor of Surgery in the department of Orthopaedic Surgery at McGill University. He is the Chief of Orthopaedic Trauma and Hand and Microvascular Surgery. He is also a co-director of the JTN Wong Labs for Bone Engineering where a large part of his research focuses on osteonecrosis and bone healing.

Elizabeth Hazel, M.D. is an Assistant Professor of Medicine in the division of Rheumatology at McGill University. She completed her Internal Medicine and Rheumatology training at McGill University. Her interests include patients with juvenile idiopathic arthritis.